

Needle-like central spikes in Rett Syndrome

Teaching Video Neuro*Images*

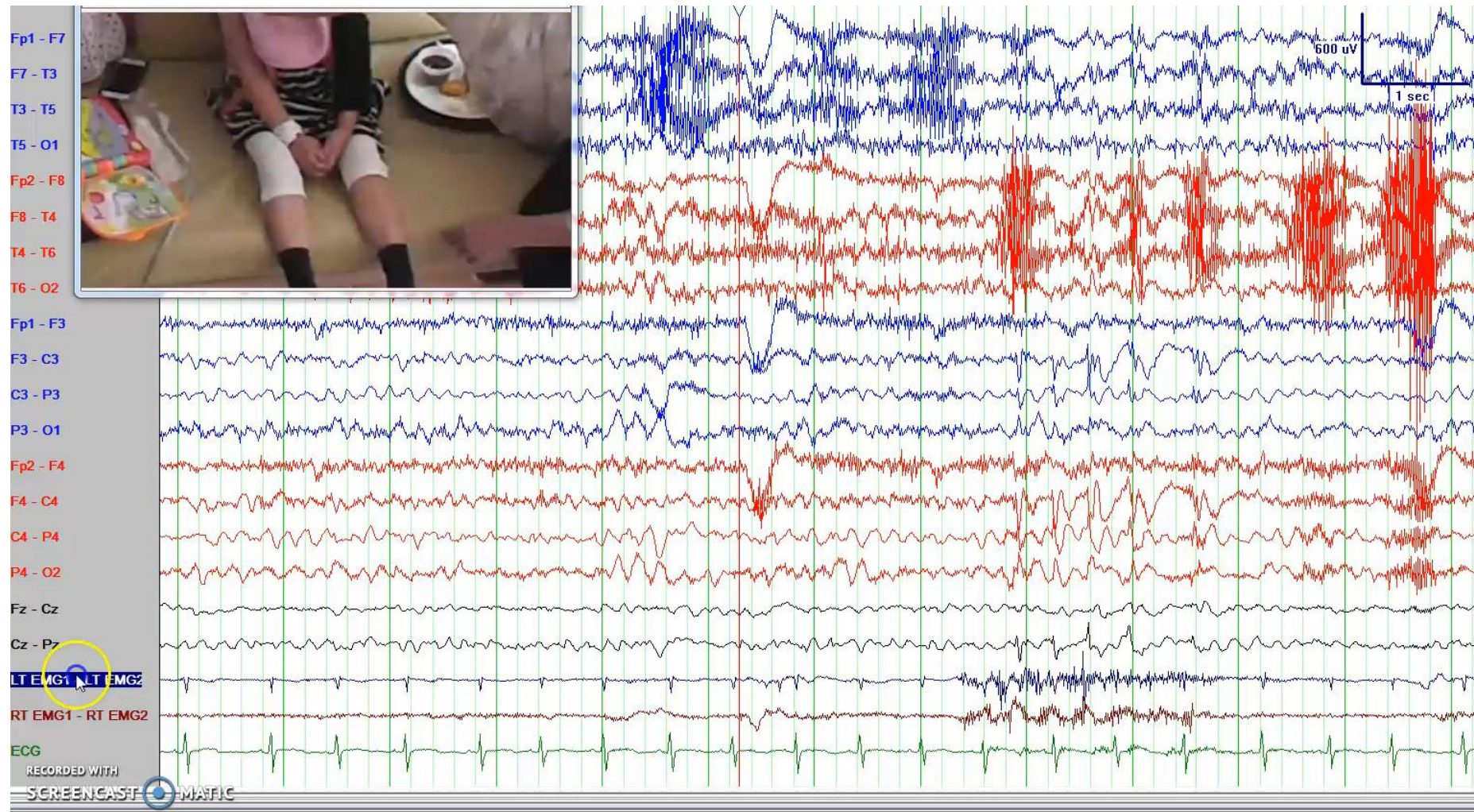
Neurology

Resident and Fellow Section

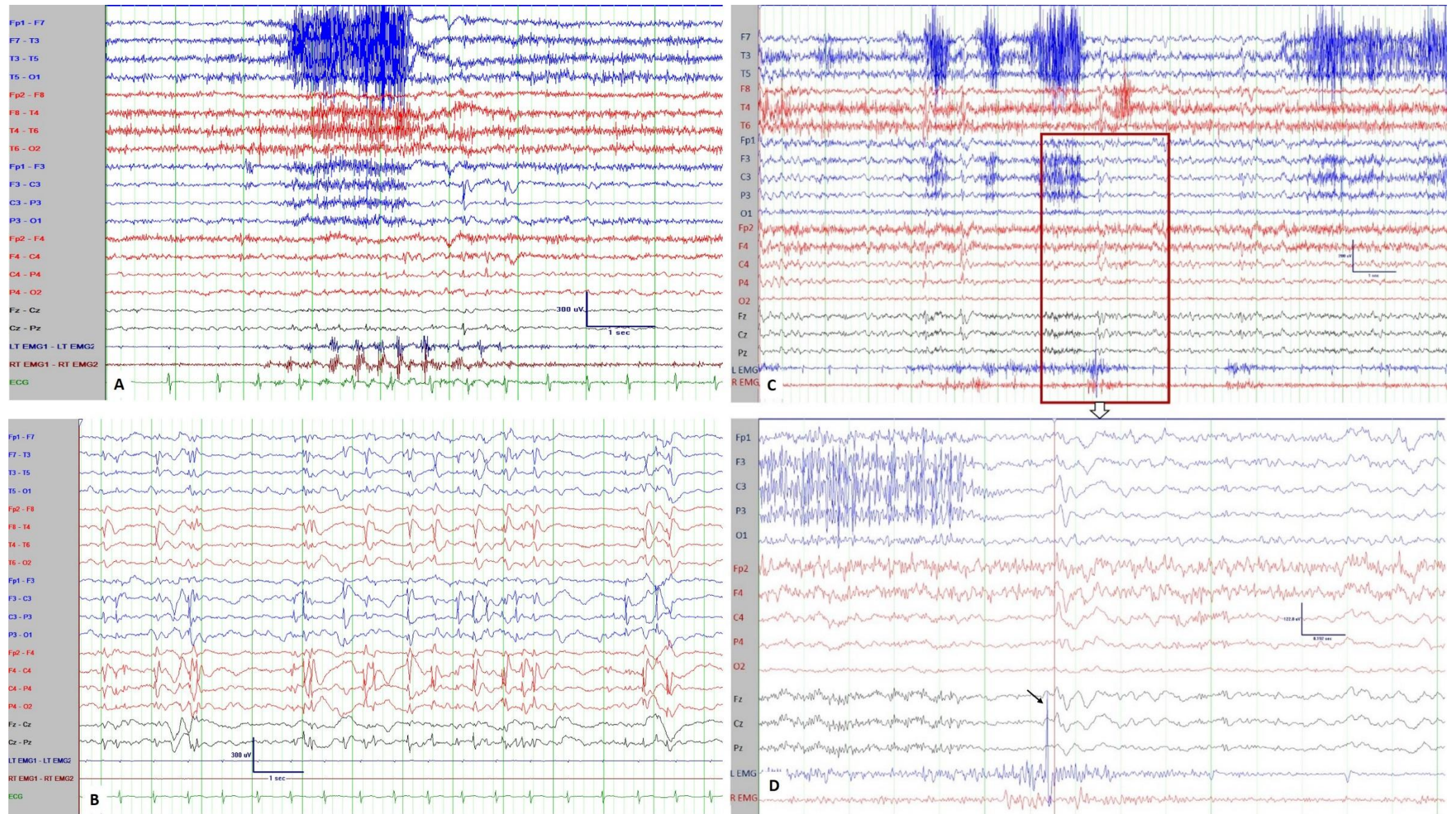
Vignette

- Four-year-old-girl with Rett syndrome (RS) presented with drug-resistant epilepsy since 4-years of age
- Her video-EEG captured rhythmic hand-tapping with bilateral central spike-and-waves

Video



Figure



Needle-like central spikes evoked by hand tapping in Rett Syndrome

- Characteristic EEG features in RS: Prominent central rhythmic theta-activity during wakefulness & needle-like central-spikes evoked by contralateral passive finger-tapping
- These central-spikes may represent exaggerated somatosensory evoked-potential secondary to the presence of hyper-excitable motor & sensory cortices
- Reflex seizures (focal and atonic) provoked by rhythmic hand-pressure have been described in two children with RS¹
- In contrast, hand-clapping has also been reported to suppress continuous centro-temporal spikes²
- Thus, the clinical significance of these provoked central-spikes is unclear